Case Report

An Unusual Case of Fever of Unknown Origin

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ABSTRACT

Takayasu arteritis is a large-vessel vasculitis that mainly affects aorta and its major branches. It most commonly affects women of 20-40 years of age. A few patients presents after an age of 40 years. Arm claudication is the most common presenting symptom, though atypical features are not unusual. A 45 year old female comes to emergency with two days high grade fever. Initially she has low grade fever, remittent in nature but not being associated with cough, chills and rigor or rash. Detailed clinical and laboratory examinations reveal absence of pulse in left upper extremity with carotid bruit bilaterally and significant raised erythrocyte sedimentation rate(ESR) and C-reactive protein(CRP) with nearly complete luminal obstruction of left subclavian artery on CT angiography mainly. Her symptoms improve on steroid therapy. Takayasu arteritis may present only with features of fever of unknown origin (FUO) alone without any significant vascular symptom and it should be considered among differential diagnoses of FUO.

Key words: Takayasu arteritis; fever of unknown origin; Angiography

INTRODUCTION

Takayasu arteritis(TA) is an inflammatory and stenotic disease, that mainly affects aorta and its major branches.[1] It is an uncommon disease with an estimated annual incidence rate of 1.2–2.6 cases per million. It is most prevalent in adolescent girls and young women. Subclavian artery is the most frequently involved artery (93%).[1] Arm claudication is the most common presenting symptom (35%)[1], but it may present with atypical features also. Here we are presenting a case of takayasu arteritis with FUO as sole clinical symptom. American College of Rheumatology Classification Criteria for the diagnosis of TA is listed Table 1.

CASE REPORT

A 45-years old female presented in emergency ward with chief complaint of fever for last 1 month. Fever was of low grade in intensity, remittent in nature, not associated with chill & rigor, cough, burning micturition, pain abdomen or rash. For the last 2 days, intensity of fever

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Received: 07 November 2020
Revised: 07 December 2020
Accepted: 12 December 2020
Published online: 01 January 2021
Citation: Mishra A, Molla K, Gantait K. An Unusual Case of Fever of Unknown Origin. J West Bengal Univ Health Sci. 2021; 1(3):76-79.

Table 1: American College of Rheumatology Classification Criteria for Takayasu's Arteritis[2]

1.	Onset before age 40 year
2.	Limb claudication
3.	Decreased brachial artery pulse
4.	Unequal arm blood pressure (> 10 mmHg)
5.	Subclavian or aortic bruit
6.	Angiographic evidence of narrowing or occlusion of aorta or its primary branches, or large limb arteries

The presence of three or more of the six criteria is sensitive (91%) and specific (98%) for the diagnosis of Takayasu's arteritis.

increased which necessitated the admission of the patient in emergency ward. There were no other complaints. Quick physical examination revealed body temperature of 102° F, pulse rate of 106 beats per min, feeble and blood pressure 96/70 mm Hg in right hand. Moderate degree of pallor was also noted clinically. Cardiac and chest auscultation were unremarkable.

In emergency ward, all obligatory investigations for FUO were performed. Significant findings were Hb-9.2 gm%, ESR: 93 mm/1st hr, CRP: 19.25 mg/lit. Other important normal findings were TLC: 6,300/ cmm, blood and urine culture: no growth, alkaline phosphatase: 125 IU/ lit, lactate dehydrogenase: 154 U/lit, CPK: 121 U/lit, creatinine: 0.9 mg, Mantoux test: negative. Chest X ray and serum protein electrophoresis both are unremarkable.

Considering it a case of FUO, patient was shifted to academic ward for meticulous examination to find out potential diagnostic clue (PDC). On detailed examination, it was found that pulse was not palpable in left brachial, radial arteries. Pulse was feeble in both carotid arteries and right radial artery. Blood pressure was not measurable in left brachial artery. On auscultation, bruit was heard in left carotid artery and left interscapular region. Though patient's age was 45 years, involvement of both carotid and subclavian arteries in form of decreased pulse, unequal arm blood pressure and carotid bruit and significantly raised inflammatory markers like ESR and CRP made us to think about Takayasu arteritis.

Then we performed CT Angiography for detailed evaluation of vascular system. CT angiography revealed severe concentric thickening with luminal stenosis (up to 90% area) of right common carotid artery. Right subclavian artery showed 70% stenosis. Left common carotid artery at lower third of neck showed mild concentric thickening with luminal narrowing (up to 40% area involvement) and there was near total obstruction of left subclavial artery. Multiple collaterals were also visible around neck and thorax (Fig 1). Renal arteries lumens were apparently normal (Fig.2).

Patient was then put on tablet Prednisone 40 mg along with calcium, cholecalciferol, rabeprazole. Fever subsided after 4 weeks. Other constitutional symptoms also disappeared gradually. Inflammatory markers also returned to normal level after 8 weeks. Then gradual tapering of steroid was done.

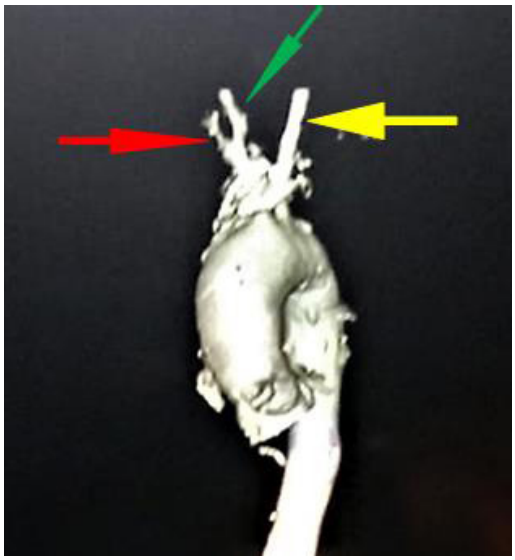

Figure 1: CT Angiography images, (Red arrow shows Right subclavian artery stenosis, Green arrow shows stenosis of Right common carotid artery and Yellow arrow shows Left common carotid artery narrowing)

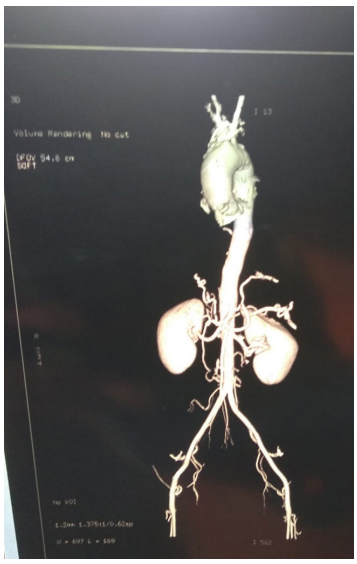

Figure 2: CT angiography- renal arteries

DISCUSSION

Takayasu arteritis is a large-vessel vasculitis that mainly affects aorta and its major branches. It most commonly affects women of 20-40 years of age; however approximately 25% of cases begin before age 20, and 10% to 20% of patients are seen after age 40 years.[1]

Takayasu arteritis is a systemic disease with generalized as well as vascular symptoms. Vascular symptoms are organ specific ranging from limb claudication (most common-35%), visual changes, syncope, abdominal pain, nausea, vomiting to atypical chest pain.[1] Common generalized symptoms are malaise (most common-20%), night sweats, myalgia, arthralgia, anorexia and weight loss.[3] About one in five TA patients presents with fever and malaise, which can be accompanied by night sweats and weight loss. A few patients present with FUO alone with minimal or no signs of vascular insufficiency for weeks or months before the diagnosis of Takayasu arteritis becomes evident.

The diagnosis is confirmed by the characteristic pattern on arteriography[4], which includes irregular vessel walls, stenosis, poststenotic dilation, aneurysm formation, occlusion, and evidence of increased collateral circulation. Corticosteroids are the cornerstone of treatment of active TA.[5,6]

Unfortunately, medical therapy can reduce inflammatory symptoms, but rarely reduces or reverses stenotic lesions. Treating stenotic lesions may require revascularization procedures[1,7,8] (Bypass surgery has better outcome than Angioplasty[6]).

CONCLUSION

Takayasu arteritis may present only with features of FUO alone without any significant vascular symptom and it should be considered among differential diagnoses of FUO. Though it is a disease of young woman, it may present even after 40 years of age. Not all the cases need vascular surgery

and steroid therapy quickly alleviates its symptoms .

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